TOWARD THE DIAGNOSIS OF CONGENITAL HEART DISEASE

by

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THE HEART IS THE TOUGHEST STRONGEST MOST PERSISTENT AND TENACIOUS ORGAN AND CARRIES ON AND ADAPTS ITSELF AGAINST ALMOST ANY ODDS OFTEN FOR A TIME WITHOUT SUPPORT OF OTHER ORGANS "

DR GORDON MURRAY

— ALPHA —

INTRODUCTION

Many congenital diseases are not fully or correctly diag nosed until they come face to-face with the cardiac surgical team performing the surgery Since the cardiac surgical field has expanded over the past decade the incidence of incorrect clinical diagnoses has decreased amazingly The Cardiologist, the Paediatrician the General Practitioner and the Cardiac Surgeon through team work have caused these favorable diagnostic results There is no reason why a 95% diagnostic accuracy pre-operatively cannot be attained in the future

Doctors are unable to look up the diagnosis as they can the treatment of the disease once the diagnosis has been made We must make the diagnosis from the symptoms and the signs presented by the patient In the case of an infant it is much more difficult for obvious reasons

The diagnosis of a disease being the most important aspect of the practice of medicine or surgery it is tantamount that we learn the signs and the symptoms which are char acteristic of each disease After much logical consideration we should arrive at the correct diagnosis

It is important in the diagnostic field that we be aware of all the other diagnostic possibilities If we are not aware of a disease or defect how then are we able to consider it in the diagnosis? It is what we do not know that is often more important than what we do know

The education of a doctor should never cease Once we cease learning the new and much of the old we soon become lost and wade blindly through the mire of despondency and

ignorance We must keep abreast of the advances in medicine through travelling reading, practical experience and carrying out research whenever possible The doctor should always be a student of his art

In the following pages the student will find much common knowledge and perhaps some which is new Due to recent advances and greater interest in cardiovascular lesions it behooves us to be able to help these little patients who are brought for help and information regarding their particular ailment

The present day public is too well informed through the medium of the press to accept many cloaked answers from their doctors as Just one of those things "He or she will grow out of it or that Nothing can be done They want to know and rightly so and they will find out what the trouble is and that something can be done It is our ethical duty to tell them correctly at the beginning if possible Too many neurotics have been made through the wrong diagnosis or misinformation or no information at all

More mistakes are made by not knowing or by not looking than by knowing and not telling or using this knowledge There is nothing worse than ignorance in motion If our diagnosis is correct then our treatment should be of benefit It is then that the patients are happiest and a better public relationship exists There is very little magic" left in medicine—only cold facts We should examine and re examine until we are sure of our diagnosis or diagnoses In medicine we should never take anything for granted but study and screen each patient carefully and follow the Golden Rule in so doing

There may be exceptions to many rules but with an open mind and logical thinking each statement as listed within these covers may be found to be steeped in greater information At times all we need in a diagnosis is a "lead" There are many leads" if we but know of them

The following arrangements classifications and abbrevi

ated statements on congenital heart disease are not original but are gathered from many authoritative and long experienced cardiologists and cardiac surgeons throughout the world The author has attempted to collect a bouquet of other mens flowers and only the ribbon with which they are bound belongs to him These are arranged for possible easy reference because voluminous works too frequently obscure good judgment

In conclusion I wish to extend my sincere gratitude to the following for their instruction and early encouragement to me in the field of thoracic and cardiac surgery Professor Clarence Crafoord of Stockholm Sir Russell Brock Sir Price C Thomas and Mr Holmes Sellors of London Professor Erik Husfeldt of Copenhagen Mr George Mason of Newcastle and Professor P R Allison of Oxford To Dr Alfred Blalock Dr Richard Overholt Dr Robert Gross Dr Charles Bailey Dr Claude Beck Dr George C Griffith and Dr Henry Swan of the United States To Dr W G Bigelow of Toronto Dr Pere Rendondo-Ramerez of Mexico City and many others from whom I have learned much through my association with them

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TOWARD THE DIAGNOSIS OF CONGENITAL HEART DISEASE

CODE FOR ABBREVIATIONS

A P W—Aortic-Pulmonary Window
A V—Arterio Venous
A V Communis—Atrio Ventricular
A A V R —Aortic Arch Vascular Ring
B B B —Bundle Branch Block (Rt or Lt)
C A or C of A —Coarctation of Aorta
C A —When associated with topic means Cardiac Arrest
D D —Differential Diagnosis
E C G —Electrocardiogram
F O or P F O —Patent Fossa Ovalis
I A S D or A S D —Interauricular Septal Defect
I V S D or V S D —Interventricular Septal Defect
I V C —Inferior Vena Cava
I C S —Intercostal Space
L V —Left Ventricle
L A —Left Auricle
M V —Mitral Valve
M S —Mitral Stenosis
M I —Mitral Insuffeciency
P A —Pulmonary Artery
P S —Pulmonary Stenosis
P P S —Pure Pulmonary Stenosis
P D A —Patient Ductus Arteriosus
R O A —Right Oblique Anterior
R A —Right Auricle
R V —Right Ventricle
S.A S —Sub Aortic Stenosis
S V C —Superior Vena Cava
S A B E —Sub Acute Bacterial Endocarditis
T A —Tricuspid Atresia
T V —Tricuspid Valve
T of F —Tetralogy of Fallot
T G V —Transposition of Great Vessels
V A—Venous to Artery
V —Ventricle Rt. or Lt

MEANS OF DIAGNOSING HEART DISEASE

The following is a guide which if followed should bring the diagnosis to those who seek it

1 Know the embryology anatomy biochemistry and the physiopathology of heart disease
2 Know of all closely similar lesions which may exist
3 Possess a good diagnostic acuity and be logical
4 A complete history of the patient and two or three examinations in a quiet well lighted room If the patient is a child or an infant then a mild sedative will bring rich rewards
5 Examine before and after exercise and babies when asleep if possible
6 Position the patient in order to bring out sounds and murmurs to the advantage of the examiner E g left lateral recumbent position for mitral disease
7 Use fluoroscopy and x rays in the most advantageous positions One examination is often not sufficient A lateral film should always be taken and other positions where indicated
8 The E C G and phonocardiograms
9 The C B C haematocrit sedimentation rate and blood cultures
10 Angiocardiograms arteriograms oesophagograms venograms and venous pressures
11 Catheterization for pressures and O saturation in the left and right chambers and great vessels
12 Circulation time, dilution dye tests and oximetry (ear)
13 Use good clinical judgment in all aspects of differential diagnosis

SOME GENERAL DIAGNOSTIC PRINCIPLES

(Congenital and Acquired Lesions)

1 Central cyanosis indicates a right to left shunt (V A)

2 Central cyanosis is best seen in the tongue and the mucous membranes Clubbing of the digits is frequently present

3 Cyanosed nails in the newborn is in keeping with central cyanosis which means an opening between both sides of the heart and something else which raised the pressure in the right side above that of the left E g Pulmonary stenosis Pulmonary hypertension Atelectasis

4 In central cyanosis and diminished pulmonary flow patients may be helped through operation

5 A diminished or increased blood flow to the lungs can be recognized by x ray fluoroscopy and the circulation time

6 Peripheral cyanosis does not indicate heart disease per se though it is common with mitral and pulmonary stenosis and equally in many normal children on a cold day

7 Congestive heart failure rarely occurs in childhood in the natural course

8 If the liver is enlarged in the presence of a gallop rhythm and cyanosis it is not apt to be Tetralogy of Fallot (T of F)

9 "A small heart is a normal heart" (unless compressed)

10 Convulsions may be due to anoxia due to a shunt causing a low pulmonary pressure as seen in pulmonary stenosis (P S)

11 An interventricular septal defect (I V S D) is the com monest cause of heart failure in children However cases of P D A and C A should not be overlooked in the diagnosis

12 If the B P is elevated in the arms then check the lower limbs for coarctation of the aorta (C of A) A throm bosis in any artery of the body will cause hypertension in varying degrees E g Pulseless disease

13 Congestive failure in infancy is initially characterized by rapid respirations liver enlargement and rales

14 The blood pressure should always be obtained in all limbs in congenital heart disease and in all cases with hypertension

15 Questions concerning x rays of the heart
 (a) Which chambers are enlarged?
 (b) Which great vessels are enlarged and are both pres ent normal and in normal position? Are their branches normal? An R O A film shows the pul monary artery enlargement best
 (c) Is the systemic venous return normal?
 (d) Is the pulmonary venous return normal?

16 The presence of an atypical continuous murmur should always raise the possibility of a complete absence of the pulmonary arteries (Blood may pass via the bronchial arteries)

17 A narrow great vessels shadow may indicate a hypo plastic aorta E g (a) A V communis
 (b) Complete transposition of the great vessels The shadow is due to the two vessels lying behind the other in a P A " view

18 Anginal pain in infants may be due to anomalous origin of the left coronary artery

19 In cerebral anoxia with convulsions in a baby who has slow respirations where clinically rapid ones would be expected the aorta most certainly arises from the R V

or is receiving large amounts of venous blood therefrom

20 Low voltage in the E C G may mean that the coronaries receive venous blood and an anoxic state exists

21 Pulseless disease is due to local arteritis with secondary thrombosis of the vessels arising from the aortic arch characterized by syncope amnesia black outs, paresis and absent pulse in these vessels It is found more frequently in the adolescent female but thrombosis oc curs in old age from arteriosclerosis at the same sites

22 A wide pulse pressure suggests P D A aortic insuffi ciency A V aneurysm heart block hyperthyroidism, arteriosclerosis of the aorta or an aortic pulmonary win dow or fistula

23 A narrow pulse pressure suggests shock aortic stenosis myocardial weakness cardiac tamponade or constrictive pericarditis

24 In persistent tachycardia check with the E C G It may not be from hyperthyroidism anxiety or infarction!

25 A large heart is an abnormal heart DO NOT mistake the wide compressed heart of pectus excavatum or kyphoscoliosis for an enlarged heart (See a lateral film of the heart)

26 Diastolic murmurs are usually organic In congenital heart disease an apical diastolic murmur does not imply organic mitral valve disease even though organic heart disease exists

27 The E C G for coronary disease should be done follow ing or during exercise only if the heart is normal at rest

28 Dyspnoea is the chief symptom of heart disease and psychosis but may arise from pulmonary disease anae mia acidosis disease of the nerves, brain and the muscles of respiration If the heart size is normal there is apt to be a pulmonary cause for the dyspnoea One should exclude the hyperventilation syndrome

29 Cough is not often due to heart disease unless the lungs

are involved secondarily E g Pulmonary hypertension pulmonary oedema or emboli

30 Pulmonary hypertension may cause pain similar to that of coronary disease but more frequently the hypertension is associated with dyspnoea asthma, emphysema V S D P D A A S D and mitral stenosis should be ruled out as a cause of pulmonary hypertension

31 Splenic colon flexure pain slipping costal cartilages and Tietzie s disease may simulate coronary pain

32 Examine the apical area in the left lateral recumbent position for best results

33 After the age of 50 calcific aortic disease is common

34 When all the pulmonary veins drain into the right auricle, then the lungs are congested The right auricle is greatly enlarged

35 A V shunts are found in the following
 (a) Anomalous pulmonary veins
 (b) I A S D
 (c) P F Ovale (large)
 (d) A V ventricularis communis
 (e) I V S D
 (f) Aortic septal defect (Aortic pulmonary fistula)
 (g) Overriding pulmonary artery
 (h) Coronary sinus and truncus arteriosus
 (i) P D A
 (j) Ruptured sinus of Valsalva
 (k) A V aneurysm of the lung (Some consider this a V A shunt)
 (l) Traumatic and inflammatory lesions and new growths rupturing into an artery and vein

36 Cerebral anoxia in the infant or child may show
 (a) Slow breathing (apnoea)
 (b) Unconscious spells
 (c) Later the child may squat to overcome the spells

as is seen in Tetralogy of Fallot and other right to left shunts

37 The absence of a murmur in a cyanosed child raises the question of complete pulmonary atresia (Isolated P S with or without I A S D always has a murmur and is always best heard in the back)

38 An unexplained fever of a week s duration is sufficient grounds for suspecting bacterial endocarditis in a patient with known valvular or septal disease

39 P D A (Patent Ductus Arteriosus) may have a systolic murmur only in early life

40 Lest we forget pericarditis look for it more often The right pericardio phrenic angle is frequently observed to be acute in x ray films

41 Pulmonary emboli are often overlooked They are com mon post operatively They may cause pleuritic pain only or be lethal due to the large size of the embolus

42 Use nitroglycerine just prior to exertion to test the pres ence of coronary disease Coronary dilatation may be come irreversible with prolonged use of nitroglycerine causing thrombosis due to slowing of the blood serum

43 Haemoptysis may arise from pulmonary hypertension One may note frequent chest colds" from A V shunts—many times mistaken for "bronchitis"

44 Leg cramps coldness numbness and non healing ulcers may be due to coarctation of the aorta (C of A) or similar arterial blockage Do not mistake for Buerger s or LeRiche s disease

45 Look for a congenital heart defect in a "squatting" child Children may also entwine their legs or lean on their laps This is more often associated with Tetralogy of Fallot

46 Pulmonary stenosis causes dyspnoea in a child It is best seen on a well populated school ground Watch the child who lags behind or who is squatting

47 Dysphagia and stridor in an infant may be due to a vascular ring about the trachea and oesophagus to an abnormal subclavian artery (Vasa lusoria), to severe pectus excavatum or an enlarged thymus

48 Increased venous pressure is found in pericarditis tamponade mediastinal tumors right heart failure, vena cava syndrome Ebstein s disease and tricuspid stenosis

49 Look to the lungs and mediastinum in obstruction of the vena cava (superior)

50 Lower body cyanosis in a child with clubbed toes may be found in infantile type of coarctation of the aorta (C of A) In adults these findings suggest a P D A with a reverse flow

51 "Bird like movements of patients are seen in thyrotoxic heart disease

52 Alcoholics suffer from vitamin deficiency and may have enlarged hearts (abnormal)

53 Splinter hemorrhages may be seen in the nails and one may find a Grade II murmur present with bacterial endo carditis

54 Bradycardia is frequently present in hypothyroidism

55 Test the carotid body reflex carefully in all cases of tachy cardia of questionable origin

56 Syncope may arise from a sensitive carotid body Turn ing the head might drop the patient (syncope) as does a tight collar or local pressure

57 Pulmonary stenosis and I V S D cause a harsh murmur along the left sternal border A thrill is very inconstant.

58 To bring out murmurs use exercise Take the blood pres sure and E C G following exercise for better evaluation

59 A large pulmonary artery is seen in

(a) Mitral stenosis
(b) I A S D I V S D
(c) P D A and aortic pulmonary fistula
(d) Eisenmenger s syn drome
(e) Multiple pulmon ary emboli

(f) Pulmonary fibrosis
(g) Pulmonary valve stenosis (post stenotic dilation)
(h) Emphysema
(i) "Idiopathic" dilatation of the pulmonary artery

60 Rarely does one see rib notching in (C of A) below age of fifteen years

61 The pulmonary second sound is increased in pulmonary hypertension E g Mitral stenosis Eisenmenger s complex aortic pulmonary window or emboli

62 *Masqueraders of a failing heart (myocardosis) are*
(a) Gastric complaints (gas)
(b) Insomnia and dreams
(c) Fatigue
(d) Ergophobia (fear of doing usual acts)
(e) Tiredness
(f) Personality changes (Morose and tyrannical)
(g) Anorexia
(h) Many changes from the normal"

63 Increasing venous pressures are seen in the early failing heart Check the venous and arterial pressures Haemorrhoids and bowel haemorrhages may be associated with heart failure pericarditis or cirrhosis of the liver

64 Do not mistake cardiac asthma for bronchial asthma as heard in the presence of a bronchial adenoma All that wheezes is not asthma'

65 CARDIOMEGALY is seen also in
(a) Anomalous origin of the left coronary artery from the P A
(b) Familial
(c) Fibroelastosis
(d) Friedreich disease
(e) Gargolism
(f) Von Gierke s disease
(g) Rhabdomyoma
(h) Myocarditis

66 A systolic murmur at the apex may be the first physical sign of anemia hyperthyroidism or a dysfunctioning sym pathetic nervous mechanism
67 It is often wise to leave well enough alone after three score years and ten
68 Remember that the heart may not be alone or be the primary cause of the patient s complaints Examine every system carefully Post operative complications are in direct proportion to the pre operative investigation prep aration selection and diagnosis
69 Collaterals prolong life they are safety valves for a time but help in a diagnosis if observed during the ex amination Varicose veins are frequently due to cardiac obstruction and not from standing
70 About 80% of the troubles in medicine are due to blocked lumina If all the lumina are open there will be few signs and symptoms of disease E g Bowels ducts ureters valves arteries lymphatics
71 The principle function of the heart is mechanical
72 The heart is made of two pumps lying side by side each serving its own portion of the body If their streams meet other than at the pulmonary or systemic capillaries then there should be clinical evidence of a lesion
73 Supernumary coronary arteries are not infrequent. The coronary arteries and veins may be joined by a fistulous tract
74 Anoxia is a concept of pathologic physiology
75 If the bronchioles are obstructed oxygen cannot enter the blood via the alveolar sacs therefore cyanosis occurs Keep the bronchi clear Many cases of pulmonary oede ma die because of anoxia from blocked bronchi only Oxygen must have a direct passage to the alveoli to be of use
76 The hyperventilation syndrome causes many vague symp toms and signs until we understand its mechanism and

biochemistry Look for it in all anxious and nervous patients

77 Anoxia may arise from
- (a) High altitudes
- (b) Emphysema
- (c) Heart failure
- (d) Shock
- (e) Anaemia
- (f) Haemorrhage
- (g) *Oedema*
- (h) Cyanide, CO_2 and drugs
- (i) *Obstruction of O_2 or* blood flow
- (j) Cigarette smoking (The elastic tissue of the lung is destroyed)
- (k) Alveolar fibrosis (Rich *syndrome*)
- (l) A V fistula
- (m) Respiratory alkalosis

78 CO_2 narcosis is not present in hyperventilation syndrome but an excess of O_2 is present There is an excess of CO_2 in emphysematous lungs Use care with oxygen therapy in the latter

79 Cyanosis from nitrites from a chicken yard from well water nearby reduces haemoglobin in the blood Ask regarding infant water supply in cases of cyanosis in babies who live in the country and use well water

80 Bilateral atresia of the posterior nares may cause anoxia and sudden death in newborns

81 There must be five grams of reduced haemoglobin per 100 cc of blood in peripheral circulation to produce visible cyanosis

82 Factors controlling cyanosis in the Systemic Circulation are
- (a) Volume of A V shunt
- (b) *Rate of utilization of O_2*
- (c) Peripheral stasis

83 Factors associated with the Pulmonary Circulation in the presence of cyanosis are
- (a) Vital capacity
- (b) Rate of flow through the lungs
- (c) Partial pressure of O_2

(d) Congenital anomalies in the pulmonary circulation
(e) Difficulty in return of oxygenated blood to systemic circulation
(f) Failure of the blood to be oxygenated as it passes through the lungs (E g A V aneurysm)
(g) Concentration of blood (E g Polycythemia)
(h) Pulmonary disease (Rich syndrome)
(i) Inadequate circulation
(j) Drugs as morphia

84 Determine the cause of cyanosis in the malformed heart by
(a) Size of pulmonary artery (Conus)
(b) Oxygen saturation of arterial blood will demonstrate presence of reduced haemoglobin insufficient to cause "visible" cyanosis
(c) Effect of inhalation of O_2 upon O_2 saturation of arterial blood
(d) Circulation time

85 Look for clubbing in cardio-pulmonary lesions It is not infrequent in pulmonary malignancy also

86 Look for central and peripheral cyanosis

87 Aetiology of dyspnoea
1 Mechanical Impairment of Ventilation
(a) Muscular weakness E g Poliomyelitis and my asthenia gravis
(b) Skeletal fixation E g Pectus excavatum
(c) Hydrothorax pneumothorax pericardial effusion or constrictive pericarditis
(d) Diaphragmatic hernia
(e) Bronchial or tracheal obstruction
2 Impairment of Pulmonary Distensibility
(a) Pulmonary congestion
(b) Pulmonary fibrosis
(c) Alveolar fibrosis (Rich syndrome)

3 Pulmonary Insufficiency
   (a) Emphysema
   (b) Inflammation of lung parenchyma
   (c) Multiple pulmonary emboli
4 Inadequate delivery of O_2 to tissues
   (a) Arterial hypotension
   (b) Anaemia
   (c) Cardiac failure
5 Hyperventilation of Central Type as from Acidosis
   (a) Nephritis
   (b) Diabetes
   (c) Hyperthyroidism
   (d) Hyperventilation syndrome

88 There are no symptoms in heart disease which are absolutely typical and can be accepted as evidence of heart disease We must ponder and weigh each finding in order to come to a logical conclusion

89 A patient may have a severe lesion of the cardiovascular apparatus without symptoms

90 Precordial pain upon exertion is frequently associated with heart disease A check of the coronary arteries is in order

91 Sleeplessness is a common sign in cardiovascular disease and is too often overlooked Too much sedation is frequently handed out by the attendant First, find the cause

92 Electrically amplifying stethoscopes are useful and bring out tones better for those whose hearing is defective

93 For a classification of cardiovascular disease, the reader is requested to refer to LUISADA's book on *Heart* pages 164 165

94 Hoarseness may be present in lesions causing dilatation of the pulmonary artery due to compression of the recurrent laryngeal nerve

95 Pericardial effusion may cause the patient to rest on his hands and knees or lean over a pillow on his lap

96 There are dysfunctions of the heart from metabolic vitamin and endocrine disturbances
(a) Von Gierke s disease
(b) Thiamine deficiencies
(c) Fatty heart
(d) Pellagra
(e) Scurvy
(f) Rickets
(g) Hyper and hypothyroidism
(h) Addison s disease and Phenochromocytomas
(i) Thymus gland (Myasthenia gravis)
(j) Hyperparathydroidism

97 Always test for postural hypotension in weak spells Take the blood pressure lying and standing Do not over look petit mal

98 Lest we forget periarteritis nodosa—a biopsy of the muscle should verify its presence

99 The blood pressure in P D A gives a low systolic and a wide pulse pressure E g 91/45 and 98/35 after exercise The average normal pulse pressure is 67 70 mm Hg

100 Pulmonary hypertension from P D A may cause a louder murmur and a larger heart or it may become less marked and disappear in the later stages

101 We may have the following combination P D A I V S D and pulmonary hypertension

102 We may have P D A I V S D and pulmonary fibrosis (arteritis)

103 Always palpate the femorals while taking the blood pressure of the upper limbs One may detect coarctation of the aorta more frequently by doing so Always verify the pulse to the area of complaint E g Carotids in cases of dizziness or syncope

104 Hyperventilation syndrome cases require more (10%) CO_2 during anaesthesia to prevent apnoea and death There is a severe CO_2 lack in the brain

105 All physicians should be capable of doing cardiac massage and BEFORE the three minutes allowed are up be

fore brain damage takes place Oxygen must be passing into the trachea freely with controlled respirations or by other artificial means

106 Relieve coronary pain by
   (a) Increase blood supply to ischaemic muscle by vasodilators or collaterally produced circulation (operation) E g Beck s or Thompson s Hypothermia is of much benefit in the severe cases of thrombosis of the coronary arteries
   (b) Decrease the metabolites and cholesterol
   (c) Section pain fibres to the heart
   (d) Avoid nicotine and stress

107 Many patients prior to and following cardiac surgery should be seen and treated by a psychiatrist The human fear of heart lesions is tremendous

108 Angina pectoris is diagnostic of coronary disease in the absence of arteriosclerotic aortic disease (stenosis) severe anaemia, luetic coronary osteal stenosis and pulmonary hypertension

109 Some manifestations of coronary disease
   1 Angina pectoris
   2 Myocardial infarction herniation and rupture
   3 Cardiac asthma or pulmonary oedema
   4 Congestive heart disease (failure)
   5 Adams Stokes Syndrome
   6 E C G Changes
      (a) Q S T-T changes or previous infarction
      (b) A V block
      (c) R B B B (Right bundle branch block)
      (d) Low Q R S Low and inverted T wave
   7 X rays
      (a) An unexplained cardiac enlargement
      (b) Absence or reversal of pulsation
      (c) Calcified coronaries may be seen

(d) Angiograms may show blockage in the coronary vessels

8 Digestive symptoms

9 Symptoms made worse by nicotine

110 Extra-cardiac pains may have origin in the heart via the nerve supply through "pain points" E g Loose click ing costal cartilage or bronchial lesions

111 Pulmonary hypertension secondary to cardiovascular (primary) disease may be recognized by

(a) Long standing cough
(b) Persistent or inter mittent cyanosis
(c) Associated dyspnoea
(d) Presence of two-type pain (one increased by deep breaths)
(e) Right ventricular en- largement.

112 The heart is an independent organ

113 The biochemistry of the bundle of His is related to nerv ous tissue It produces acetylcholine

114 "The heart muscle is one of the unfatiguable muscles the others are bird wings in long flight"

115 A recumbent position allows for greatest diastolic filling Fowler s position is not always suitable in heart failure cases

116 A large heart frequently gives short contractions and therefore poor propulsive power However in A V block there is a normal cardiac output The propulsive power of each beat must be tremendous and the heart is very large in diastole

117 "The heart is a pump with an intricate mechanism with individual physiology of each left and right unit."

118 It is difficult to be sure of the exact site of the catheter in catheterizing the heart Errors may be present The catheter report should agree with the clinical findings

119 Pulmonary stenosis may give convulsions from cerebral anoxia

120 The importance in diagnosing disease still remains with

The history and clinical examination of the patient, fluoroscopy x rays and angiocardiograms Catheterization is used in borderline cases where verification is essential

121 Dr Forsman is "Father of Catheterization In November 1929, he was the first to carry out the procedure, and on himself

122 Speed up the machine (angiocardiograms) in children as their hearts are more rapid We may use the Weber manoeuvre to slow the heart rate in adults Medications may be used also

123 It is best to estimate the blood postassium in mitral lesions If it is rising, it is more dangerous for operation

124 Differential diagnosis

Mitral Stenosis	*Mitral Insufficiency*
(1) Early dyspnoea late fatigue	(1) Earl fatigue, late dyspnoea.
(2) R V is enlarged in presence of pulmonary hypertension	(2) L V enlarged
	(3) Systolic murmur
	(4) Normal P A
(3) Diastolic murmur at apex	(5) Pulmonary congestion
	(6) Less calcium in the valve
(4) Large P.A	
(5) Pulmonary congestion	(7) L A. less enlarged
(6) Calcium deposits in the valve	
(7) L A enlarged	

125 Ballistocardiograms might have a place in the diagnostic setup in the future of heart disease

126 Be on the lookout for cardiac neurosis but do not make a patient a neurotic through a wrong diagnosis or misplaced treatment.

127 Nitroglycerine relieves the pain of coronary disease but does not cure the patient Certain operative procedures are found to give relief of symptoms due to formation

of new collaterals to the ischaemic sites

128 Diabetes nicotine and coronary disease join hands fre quently and "pass out together

129 Pulmonary embolus think of it for it is common! Aeti ology Phlebitis somewhere (post op) Evidenced by
   (a) Acute pleuritic pain
   (b) Cough and rales at the site of lodgement
   (c) Low grade fever
   (d) Haemoptosis and dyspnoea
   (e) X ray shows triangular and dome lung shadow more often at the bases

130 Lest we forget! Obstruction of the S V C causes a def inite syndrome Venous pressures and venograms are of value here If both S V C and I V C are obstructed, think of constrictive pericarditis

131 There are cardiac tumors cysts and aneurysms which simulate other cardiac lesions Is it actually the heart at fault?

132 A dissecting aneurysm is too frequently forgotten in an early diagnosis

133 Generally we die from cardiovascular disease Cancer is now fourth on the list of causes of death

134 Regional hypertension is not only seen in coarctation of the aorta but arterial thrombosis as well E g Aortic arch syndrome compression of the aorta or arteries by tumors horseshoe kidney tumors of arteries themselves and fibrosing lesions of the arteries

135 Consider the heart as two separate units—one supplying the lungs the other supplying the body generally The two units unite only through the alveoli and peripheral arterio venuoles If other connections are present then an abnormality exists

136 The cardiac silhouette is frequently altered by hyper ventilation partial or complete bronchial obstruction and by cyst of the lung or congenital heart disease

CLINICAL DIAGNOSIS OF HEART DISEASE IN CHILDREN

(*Congenital and Acquired*)

(Patterson)

"EYES FIRST AND MUCH"

A CYANOSIS

Causes (1) Methaemoglobinemia
(2) Cerebral
(3) Pulmonary Decreased on crying, responds to O_2
(4) Cardiac Increased on crying indicates right to left shunt Does not respond to O_2
(5) Arteriovenous mixing of blood Eg Pul A V fistula

B DYSPNOEA

(1) Is the breathing more rapid than normal?
(2) If it always breathes faster" without stridor or cyanosis, then it is apt to be left heart failure with pulmonary congestion or pulmonary stenosis with decreased pulmonary blood flow Check the red blood count haematocrit and look for diabetes salicylates poisons or fever

C CARDIAC ENLARGEMENT

(1) If more than 1 cm beyond mid clavicular line it indicates enlargement Check the E C G, position of the diaphragm and expiration for atelectasis pectus excavatum pneumothorax pulmonary hypertension pericardial effusion or agenesis of a lobe or lung

(2) If the palpating finger is lifted at the apex it is apt to be left ventricular hypertrophy If the finger is lifted" over the pulmonary artery area it is apt to be pulmonary hyper tension (Right ventricular hypertension)

(3) Anaemia malnutrition myocarditis hyper thyroid endocardiofibroelastosis glycogen storage disease coarctation or medial arterio-sclerosis are other causes of enlargement

D TACHYCARDIA

(1) If the pulse is above 110 per minute when asleep it suggests myocardial disease or hyper thyroidism

E BLOOD PRESSURE

Always check for (using both arms and legs in C A suspect)

(1) Coarctation of the aorta (C A)
(2) Phenochromocytoma
(3) Wide pulse pressure is present in P D A and aortic window
(4) A narrow pulse pressure is present in aortic stenosis

F RIGHT HEART FAILURE

Note (1) *Oedema* (3) Neck veins filling
(2) Hepatomegaly

G MURMURS

Note (1) Character (4) Position
(2) Timing (5) Transmission
(3) Quality (6) Thrill

In children give sedation (1/10 gr barbiturate/lb body weight rectally)

H CATHETERIZATION ANGIOCARDIOGRAMS X RAYS AND PHONOCARDIOGRAMS

These are other means of diagnosing congenital heart lesions

I S A B E

Note (1) Blood cultures (2) Emboli in skin

J Collagen Diseases

E g (1) Lupus erythematosus
(2) Polyarteritis nodosa
(3) Rheumatoid arthritis
(4) Dermatomyositis
(5) Disseminated scleroderma

K Anaemias

Check the blood

L Myasthenia Gravis

Check by giving prostigmine

M Avitaminosis

Check the diet

FOETAL ANOXIA AND CYANOSIS IN THE NEWBORN

Anoxia in the new born is either from poor oxygenation or from a circulatory shunt which prevents oxygenated blood from reaching the tissues

INTRAUTERINE ASPHYXIA

The foetal organ of respiration is the placenta

It depends upon

(a) Adequate supply of oxygen in the maternal blood
(b) Intact placental structure and circulation
(c) Intact pathways for blood from the placenta to the foetus

These interfere with foetal oxygenation

(a) Maternal disease
(b) Placental disease
(c) Compression of cord
(d) Atelectasis
(e) Functional depression of respiratory centre from mother s drugs
(f) Endothelial fibroelastosis

Respiratory distress (Cyanosis) in the newborn (other than congenital heart lesions) may be caused from

(1) Atelectasis
(2) Hyaline membrane disease
(3) Alveolar dysplasia
(4) Tension pneumothorax or cysts
(5) Congenital cysts of the lung
(6) Aortic rings Vasa lusoria
(7) Tracheo-oesophageal fistula
(8) Diaphragmatic hernia
(9) Bilateral atresia of the posterior nares
(10) Microglossia

(11) Tetany
(12) Myasthenia gravis
(13) Intracranial hemorrhage
(14) Agenesis of lobe or lung
(15) Severe pectus excavatum

"Anoxia not only stops the machine but wrecks the machinery"

HALDANE

THE CONGENITAL HEART DISEASES
A CLASSIFICATION

(Paul Wood)

ACYANOTIC

A No Shunt

1 *Simple*

(a) Dextrocardia

(b) Congenital hypertrophy

(c) Heart block

2 *Left Hypertrophy*

(a) Coarctation of the aorta (C A)

(b) Congenital aortic stenosis

(i) Subvalvular (ii) Valvular

(c) Bicuspid or quadricuspid valves

(d) Bicuspid plus aortic incompetence

3 *Right Hypertrophy*

(a) Pure pulmonary stenosis

(i) Valvular (ii) Infundibular

(b) Mild pulmonary sternosis with V S D or A.S D or P F Ovale

(c) Pulmonary valve incompetence from congested dilated P A (Pulmonary plethora)

B A Shunt *(Left to right)*

1 I V S D 3 P D A

2 I A S D 4 *Aortic pulmonary Fistula (Window)*

CYANOTIC

A Right to Left Shunt *(Pulmonary Oligema)*

(a) Cyanosis from birth

1 *Right Hypertrophy*
   (a) Tetralogy of Fallot
      (i) Infundibular stenosis   (ii) Valvular stenosis
   (b) Marked P V stenosis with I V S D or I A S D or P F Ovale (Triology of Fallot)
   (c) Persistent truncus
2 *Left Hypertrophy*
   (a) Pulmonary valve or artery atresia with septal defects
   (b) Tricuspid atresia with septal defects ("a and b" have non functioning right ventricle)
3 *Late Cyanosis*
   (a) Eisenmenger Syndrome
   (b) Moderate P S with septal defects

B LEFT TO RIGHT SHUNT
   1 *Transposition of Great Vessels*

ANATOMICAL DIAGNOSIS IN HEART DISEASE

WE MUST NOTE

(1) The pulse
(2) Neck veins (deep jugulars)
(3) Cyanosis
(4) The apex beat
(5) Liver and oedema
(6) Palpation of precordium
(7) Heart sounds
(8) Heart murmurs
(9) Fluoroscopy and X rays
(10) Cardioangiograms
(11) Catheterization
(12) Arterial and venous pressures

HEART

(Maurice Campbell)

CONGENITAL MALFORMATIONS

NON CYANOTIC

PULMONARY BLOOD FLOW

A *EXCESSIVE*
1 P D A
2 I A S D
3 I V S D
4 Anomalous pulmonary veins
5 Aortic pulmonary window
6 Triology of Fallot?

B *NORMAL*
1 Coarctation
2 Vascular ring
3 Aortic stenosis
4 Aberrant coronary

C *INADEQUATE*
1 Pure pulmonary stenosis

CYANOTIC

PULMONARY BLOOD FLOW

EXCESSIVE
1 Transposition of great vessels
2 Eisenmenger complex (Late)

INADEQUATE
1 Pulmonary stenosis and I A S D (Triology of Fallot)
2 Tetralogy of Fallot
3 Tricuspid Atresia
4 Ebstein's deformity

TRUE——TRUNCUS ARTERIOSUS——PSEUDO
PULMONARY ART FROM——SINGLE——PULMONARY ART FROM
MAIN CHAMBER VENTRICLE RUDIMENTARY CHAMBER

PATENT DUCTUS ARTERIOSUS (P D A)

(14% of all lesions, 1 5500 births)

1 Sex incidence 5 1 females
2 Growth stunted (45%-75% blood loss to lungs)
3 Fatigue and dyspnoea are dependent on the size of the passage
4 Pulmonary congestion so called "chest colds"
5 Epistaxis
6 No cyanosis or clubbing (until failure)
7 Corrigan pulse at the wrist
8 A machinery murmur" is found at the 2nd left I C S systolic and diastolic in time Early there may be a systolic murmur
9 A murmur at the 2nd left intercostal space (I C S) trans mitted to axilla and interscapular area A mid diastolic murmur may be heard at the apex
10 A "machinery murmur" occurs when the systemic diastolic pressure becomes greater than the diastolic pressure in the pulmonary circulation
11 The continuous murmur may be lacking and only a systolic is heard over the pulmonic area (in infants and children)
12 A thrill is found in the 2nd and 3rd intercostal space in 75% of cases
13 The 2nd heart sound is split
14 A wide pulse pressure A low diastolic
15 Cardiac enlargement and failure of the myocardium may occur later
16 Hilar dance (Pulmonary Plethora) is rarely noted in fluoroscopy

17 The pulmonary conus is enlarged in x ray
18 Prominent aortic arch near the P D A which pulsates markedly
19 A high P A pressure
20 Increased O_2 blood in the pulmonary artery
21 Aphonia or hoarseness is present if P A presses on the recurrent laryngeal nerve
22 Severe dyspnoea and palpitation when failure occurs
23 E C G Left axis deviation The E C G may be normal in 83% of cases

DIFFERENTIAL DIAGNOSIS *(Continuous murmur)* Seen in
(1) Venous hum (diastolic accentuation)
(2) Severe anaemia (usually systolic murmur)
(3) Pulmonary artery pressure up
(4) A V fistula of lung
(5) Rheumatic heart disease
(6) I A S D (systolic murmur)
(7) Rupture of sinus valsalva into right ventricle
(8) Compression of arteries E g Cervical
(9) Aortic pulmonary window
(10) Coronary artery fistula
May have reverse ductus flow—cyanosis

GENERALLY

"Any loud systolic murmur (without diastolic element) in acyanotic cases arises from a septal defect or pure pulmonary stenosis"

IN E C G

If P R interval is prolonged it is apt to be an A S D as conduction is longer One may have a partial right B B B" (95%)

In the early months the pressures between the aorta and the pulmonary artery are frequently equal Therefore there is no murmur sometimes One may have a systolic murmur only

DIFFERENTIAL DIAGNOSIS (D D) OF P D A AS TO DEGREE

1 *Small P D A*
   (a) The heart is normal in size
   (b) The cardiac impulse is not forceful
   (c) The x ray findings are minimal
   (d) The diastolic pressure is down slightly

2 *Average P D A*
   (a) The heart is enlarged The left border in the conus area is the chief site of enlargement
   (b) An intense murmur and thrill are present
   (c) The diastolic pressure is down
   (d) Pulmonary plethora (chest cold) is frequent

3 *Tremendous P D A*
   (a) The heart is large Early failure is noted
   (b) The heaving heart beat may even shake the body of the child
   (c) Forceful neck pulsations are present
   (d) Marked lung changes are due to excess blood flow and pressure
   (e) A great depression of the diastolic measurement pres ent
   (f) Additional murmurs may be heard

CLASSIFICATION OF P D A *BASED ON AUSCULATORY FINDINGS*

1 Cases with typical continuous murmurs
   (a) Without diastolic at the apex (90%)
   (b) With diastolic at the apex
2 Cases with an atypical murmur at the pulmonary area
   (a) Without diastolic murmur at the apex
      (i) No cyanosis (ii) Cyanosis with reverse flow
   (b) With diastolic murmurs at the apex
3 *Cases without murmurs at the pulmonary area*

Disease is from old and nothing about it has been changed It is we who change as we learn to recognize what was formerly imperceptible for we seek only what we look for we recognize only what we know”

CHARCOT

COARCTATION OF AORTA (C A.) (8%)

1 16 000, Males 5 1

A ADULT *(Majority) (Post ductal)*
Ductus open or closed

B INFANTILE *(Preductal)*
Ductus open or closed

1 Palpitation
2 Throbbing in head
3 Breathlessness
4 Early Fatigue
5 Nervousness (tension)
6 Headache
7 Dizziness
8 Epistaxis
9 Blurring of the vision
10 Precordial pain
11 Leg cramps (Do not mistake these for Leriche's disease)
12 Cold feet Warm hands
13 Tinnitus
14 Gnawing shoulder pains and throbbing carotid arteries
15 Chest and back pain is due to arterial pressure
16 Hypertension in the upper limbs
17 Hypotension in the lower limbs or no pressure
18 Decreased abdominal pulse or it is absent
19 Palpable vessels in the scapular areas
20 Collaterals are increased
21 The upper body is well developed
22 Cyanosis is not present except in preductal type of C A and seen in the legs
23 Leg ulcers may be pres ent
24 Systolic murmur (inter scapular) is heard along the left posterior chest
25 Hypertrophy of the left ventricle
26 The ascending aorta is dilated
27 Absent aortic knob (Figure "3") is noted at the knob site
28 Rib notching (Rare be fore 15 years)

29 Left axis deviation

30 Cardiac failure may occur early Apex out and down to left

31 P D A is associated with C A (In 5% of cases)

32 Bicuspid aortic valves (42%) are present with infantile C A

33 A berry aneurysm may be present in the circle of Willis This may rupture and cause death

34 Rupture of cerebral arteries show 50% of deaths

35 Normally B P is higher in legs than in arms

36 There may be a double stenosis

37 Associated with C.A there may be

(a) Bicuspid valve (9%)
(b) Aortic stenosis (10%)
(c) P D A (5%)
(d) Mitral stenosis (5%)
(e) Intermittent claudication (8%)

38 The optimal age for operation is the second decade (8 12 years) Earlier if severe

39 In 22% of cases there is aortic rupture from coarctation

40 A left S V C with coarctation and superficial collaterals have been noted on occasion

41 There may be post stenotic dilatation in C of A

42 Coarctation may occur at any site Check the B P at various levels as well as intra arterial pressures and aortograms

43 Murmurs and thrills in unusual places may be caused by C A

44 Eighty per cent die before 35 years of age

45 The majority are males

46 High blood pressure in the right arm and low pressure in the left arm is caused by

(a) Atresia of the left subclavian artery
(b) Stenosis of the left subclavian artery
(c) Coarctation proximal to the left subclavian causing pressure in the S artery

47 High blood pressure of the left arm and low blood pressure of the right arm may be from
(a) Stenosis right of the subclavian artery
(b) Origin of the right subclavian as a 4th branch of the aorta at a point beyond the coarctation
48 A hypoplastic aortic arch knob is noted in the fluroscopy or in the x ray
49 A figure "3 is noted at the C A site
50 An altered contour of the esophagus has been noted
51 A prominent left border of the superior mediastinum is *due to a dilated subclavian artery*
52 Angiograms when in doubt, should be done
53 The cardiac apex is rounded
54 The internal mamary and subclavian arteries are dilated and may be seen in the lateral film

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"We should not count our words but make our words count"

AORTIC ARCH VASCULAR RINGS

(A A V R 1 250 000)

There are many vascular malformations in the superior mediastinum Many do not cause clinical symptoms

Among those anomalies which cause symptoms and signs are the following

1 Double aortic arch
2 Right aortic arch with a left ligamentum arteriosum
3 Anomalous innominate artery
4 Anomalous left carotid artery
5 Aberrant right subclavian artery

Double Aortic Arch Features

1 The trachea and oesophagus are constricted There is stridor and dysphagia
2 Increased respiratory rate is noted The baby struggles for breath and regurgitates its food The symptoms are aggravated by fluids
3 There may be intercostal and subcostal indrawing
4 A loud wheeze can be heard with the stethoscope
5 Cyanosis is frequent in more constricted cases
6 The infant lies with the head hyperextended
7 The x ray tracheogram bronchoscopy and/or angiograms will visualize the lesion present
8 A blowing systolic murmur is present in 20% A gallup rhythm is usually present

Right Aortic Arch with a Left Ligamentum Arteriosum

Here the ligamentum crosses on the posterior aspect of the oesophagus and trachea and therefore constricts these

structures Signs and symptoms are similar to those of a double aortic arch except for the position of the arch

Anomalous Innominate Artery

Here the artery leaves the aortic arch too far to the left and in order to reach the right cervical region must pass anterior to the trachea causing constriction of this structure with consequent signs and symptoms

Anomalous Left Common Carotid Artery

Here the artery arises more to the right on the aortic arch and in order to reach the left cervical area must constrict the trachea The signs and symptoms are those of the innominate anomaly

Aberrant Right Subclavian Artery

The artery takes off independently from the aortic arch instead of off the innominate artery It ascends and passes to the right to reach the apex of the chest It passes posterior to the oesophagus and so to the right arm

---

The heart is the unfatiguable muscle"

SUBAORTIC STENOSIS (S A S)

1 24 000

Clinical Features

1 History of "heart disease" from birth
2 No history of rheumatic fever
3 A loud mid systolic murmur (80% of cases) It is an ejection type of murmur It is more commonly lower and to the left than at the aortic area
4 Moderate left hypertrophy
5 Normal blood pressure
6 Like those found from rheumatic fever causing aortic stenosis
7 Pallor and dyspnoea
8 Apical impulse may be displaced down and outwards
9 Pulse pressure may be down
10 A thrill may be felt passing to the neck Chiefly in the 2nd right intercostal space (I C S)
11 Pulsation of a dilated sinus of Valsava may be seen in a R O A view on screening

ATRESIA OF AORTIC ORIFICE

(Functional two-chambered heart)

1 39,000

(Julius Tandler 1869 1936)

FINDINGS

1 P D A and P F ovale are present
2 The right atria the right ventricle and the pulmonary artery are enlarged
3 The left ventricle is thick walled and a small chamber is noted

RIGHT AORTIC SINUS ANEURYSM
-VALSALVA-

A CONGENITAL (44 cases reported) B ACQUIRED

Associated with coarctation S A B E syphilis and aortic regurgitation

1. Tend to rupture into the right heart
2. Severe overloading of the right cardiac chambers when the rupture occurs
3. Sudden dyspnoea
4. A humming murmur

AORTIC-PULMONARY WINDOW (FISTULA)

(Aortic septal defect)

1 It is an incomplete division of the truncus arteriosus
2 Often many features of P D A are noted
3 It is associated with Eisenmengers, if the opening is less than 75 cm (Acts like a P D A but the murmur is lower)
4 A loud systolic murmur is present over the precordium (Max in 3rd and 4th I C S left of the sternum)
5 Pulmonary hypertension and P A aneurysm are complications
6 Pulmonary plethora (chest cold) is frequent
7 Progressive cardiac enlargement is noted
8 There is absence of cyanosis and polycythemia
9 Cardioangiograms may show dilution of dye in P A and an indentation of dye from aortic cross flow An aortogram will show flow to P A
10 Treatment is by closure, if no irreversible damage to the P A system

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"In surgery we should do as little as possible but as much as is necessary and herein lies good judgement"

TETRALOGY OF FALLOT (T of F)

1 8,500

THE TETRALOGY CONSISTS OF

1 Aorta drains ventricles
2 Right ventricle enlarged
3 Pulmonary valve stenosis and infundibular stenosis in 99%
4 I V S D

FEATURES

1 Marked cyanosis variable May require exercise to show it
2 Dyspnoea
3 Dyspnoea increased on feeding Paroxysmal in character may be severe
4 Unconscious spells may cause convulsions from anoxia or thrombosis of the cerebral vessels may occur causing paralysis
5 Limited activity + +
6 Retarded growth + or
7 Clubbing of fingers + + +
8 A precordial bulge Right ventricle is enlarged
9 Turner s syndrome (gonads) associated (20%)
10 "Squatting" when tired Physical capacity greatly impaired
11 Deciduous dentition
12 A small peripheral pulse
13 A systolic murmur Thrill (50%) If high 1 2 it is apt to be valvular type At 2 - 3rd left space (10% have no murmurs)
14 Red blood count 6 - 7 million + + +
15 Haematocrit 60% + or
16 Pale lung fields (Oligemia)

17 Sabot shaped heart - or + Raised apex
18 Concaved pulmonary conus area
19 Pulmonary window In L A oblique x ray A small left heart is noted
20 E C G deep S" Lead i High R" in lead iii
21 Single 2nd sound at base
22 Dextroposition aorta (20%)
23 Low arterial O_2 content
24 A heavy heart action is noted following exertion
25 The pulmonary artery is small
26 Post stenotic dilatation may be seen in the pulmonary artery, in the x ray and especially at operation

OLIGAEMIC" = Light Lung Field (V to A)

PLEONAEMIC = Dense Lung Field (A to V)

INFUNDIBULAR STENOSIS is frequently present with T of F

CATHETERIZATION (Catheter) Features in T of F
1 O_2 unsaturation arterial blood
2 High pressure in the right ventricle, low in the P A
3 The pressures in the aorta, right ventricle and brachial are increased
4 Right to left shunt Shows site and presence of pulmonary stenosis
5 Decreased pulmonary flow

RECORDS (pressures) with a catheter in the pulmonary trunk (withdraw)
Note sequence
(a) A sharp rise in the systolic to the right ventricle The systolic pressure as catheter passes through the valve indicates valvular stenosis
(b) No change in the systolic pressure with a fall in diastolic pressure when the catheter passes via the valve followed by a rise in the systolic to the ventricular level

at the infundibular region indicates infundibular stenosis

(c) A combined type indicates both lesions

"We should do the greatest amount of good and the least amount of harm we should avoid iatrogenic lesions at all cost

---

PULMONARY STENOSIS WITH NORMAL AORTIC ROOT

15% Infundibular, 90% Valvular

1 The systolic pressure is the same in P A and R V If former lower than latter then stenosis exists A gradient of 10 mm is normal All degrees are found
2 In valvular stenosis, the cusps are fused with a small circular hole present
3 Infundibular stenosis is circumscribed and is situated on the borderline between the main part of the right ventricle and the infundibulum The latter forms a cavity or third ventricle"
4 The whole infundibulum may be stenosed and forms a narrow channel which extrudes as far as the pulmonary valve
5 P S is often associated with other anomalies--chiefly I A S D and I V S D
6 A large left to right shunt dilates the pulmonary artery
7 I V S D is not uncommonly combined with stenosis of the infundibulum
8 The stenosis may be so slight that the left to right shunt is still present through the defect
9 In severe stenosis a right to left shunt occurs and is more difficult to diagnose from T of F (An overriding aorta is present here)
10 In I A S D the direction of the shunt is determined partly by the resistance of the ventricle during diastole In an uncomplicated case the resistance is lower in the right ventricle than in the left ventricle Therefore the shunt is left to right With hypertrophy of the right ventricle

there is increased resistance of the right ventricle and the shunt is right to left

11 In I A S D , if the stenosis is severe there is a right to left shunt

12 In a P F O with complete functional closure the shunt can take place in one direction only i e Right to left and may cause cyanosis (T of F)

13 Types of P S with a normal aortic root

 (a) Intact V and A septa

 (b) With I V S D and a normal A septum

  (i) With left to right shunt (ii) With right to left shunt

 (c) With P F O and intact V septum

  (i) Without shunt (ii) With right to left shunt

 (d) With I A S D and intact ventricular septum (Triology of Fallot)

  (i) With left to right shunt (ii) With right to left shunt

 All the above may be confirmed

14 Pulmonary stenosis hemodynamically is characterized by the inability to maintain an adequate cardiac output and thus the right heart takes the burden Pressure increases in the right ventricle and the right auricle The more the stenosis the less the output—even at rest This is com pensated by an increase in the arteriovenous oxygen differ ence and slight peripheral cyanosis may appear

15 In septal defects with a right to left shunt the pulmon ary blood flow is still decreased further and there is arterial oxygen desaturation

16 Severe right ventricular hypertrophy may result in a sufficient rise in pressure in the right auricle to cause a right to left shunt

17 Marked cyanosis in I A S D and P S comes later in life

18 Clinical features of P S (primary)

 (a) Loud systolic murmur over P A area

(b) Increased fatiguability
(c) Dyspnoea
(d) Slight cyanosis on exertion
(e) Syncope on occasion

Physical examination reveals the following

(a) The child may be normally developed
(b) Slight cyanosis is present An atrial wave is seen in the neck veins
(c) Very occasionally the child squats—not nearly as common as in T of F
(d) Right ventricular hypertrophy
(e) Apex beat is displaced BACKWARDS and is not palpable
(f) The enlarged right ventricle strikes the chest at each heart beat causing a parasternal or substernal lift

Sounds and Murmurs in Primary P S

1 A systolic murmur over the 2nd left I C S
2 *It occurs 05 and 06 after the first sound in the phonocardiogram*
3 Pul second shows a decreased amplitude There is an increased split of the second heart sound heard in mild and moderate cases
4 In severe P S the 2nd heart sound over the pulmonary area cannot be detected (due to aortic sound)
5 In infundibular stenosis the degree of splitting of the second sound is the same as in valvular stenosis The murmur is between the 3rd and 4th I C S The pulmonary component is audible as a faint extra sound after the end of the murmur Many believe that infundibular stenosis is secondary hypertrophy of the area due to P S and not primary This however does not include the isolated" cases of infundibular stenosis

X ray Findings in P S

1 The main trunk of the pulmonary artery is dilated
2 The peripheral vessels of the lungs are narrow

3 The right ventricle is hypertrophied and rotated to the left
4 The R A may be enlarged with a prominent appendage
5 Post stenotic dilatation is a classic finding
6 The central vessels of the hilus are dilated the peripheral narrow
7 The apex of the heart is NOT turned up in P S as it is in T of F

Angiocardiography

Inject locally (Stockholm technique)
Best inject into the R V direct via catheter
A conical or dome shaped membrane may be noted

SIMPLE PULMONARY STENOSIS WITH A RIGHT TO LEFT INTERATRIAL SHUNT

1 These at first are acyanotic until teens or "twentys when the right side pressure rises giving right to left shunt Here valvotomy is correct
2 If no right to left shunt then right ventricle failure occurs earlier

T OF F DIFFERENTIAL DIAGNOSIS AND X RAY AXIOMS

1 Those with a large aortic knob and marked pulmonary concavity (10%) are generally pulmonary atresia
2 Normal heart small pulmonary "bay" (30%)
3 Those with infundibular stenosis (30%)
4 Those with right aortic arch and no concavity (30%)
5 Do right ant oblique to show conus area

INCREASED VASCULARITY OF THE LUNGS IN T OF F PRESENT IN

1 Bronchial artery hypertrophy
2 Unilateral pulmonary artery atresia (A unilateral plethora is noted)
3 A large left to right ventricular shunt without cyanosis (1 40)

DIFFERENTIAL DIAGNOSIS OF TETRALOGY OF FALLOT

1 Aortic atresia (Note heart enlargement and vascularity)
2 Transposition of great vessels (enlargement and vascularity
3 Single ventricle with pulmonary stenosis (Use Diodrast)
4 Tricuspid atresia (Note E C G)
5 Pure pulmonary stenosis (See ante)
6 Severe P S with P F ovale (Large heart large P A)
7 Eisenmenger (Less cyanosis, better tolerance large P A)
8 Persistent truncus
9 Pulmonary artery atresia
10 Marfan's syndrome
11 Pulmonary arteriovenous fistulae

SQUATTING MAY BE SEEN IN

1 Tetralogy of Fallot (Others have different cardiac contour)
2 Truncus and pseudotruncus arteriosus
3 Certain cases of tricuspid atresia
4 Single ventricle

INFUNDIBULAR STENOSIS TYPES 3RD VENTRICLE"

1 Band
2 Subvalvular
3 Intermediate
4 Tubular

"The correct diagnosis is the 'pearl' in medical practice Let us not be satisfied without it"

PURE PULMONARY VALVULAR STENOSIS (P P S) (No other defects)

1 14 000

1 Pulmonary arthropathy + if fossa ovale remains open
2 Rough systolic thrill in upper part of the chest left side Long if severe
3 Enlarged right ventricle
4 Murmur is systolic Harsh 2nd left, near sternum
5 Cyanosis late and peripheral and with patent F ovale on occasion
6 Globular heart enlargement
7 Increased shadow of main pulmonary artery
8 Progressive breathlessness on exertion
9 Anginal pains and syncopal attacks in severe cases
10 Conspicuous "a" wave in the venous pulse
11 Pulmonary oligemia Small peripheral pulmonary vessels seen in x ray
12 Post stenotic dilatation of P.A
13 Hypoplasia of aorta and left ventricle
14 E C G right axis deviation Sharp "P" wave
15 Pressure gradient of 10 15 mm Hg considered evidence of stenosis
16 If systolic pressure is below 70 mm Hg (R V) apt to be no symptoms If above 70 mm Hg then symptoms are present
17 Right ventricle "strikes" wall of chest ("A tap")
18 A precordial bulge is present
19 A negative "T" wave in V_1 V_4 indicates severe stenosis

"We see only what we look for We recognize only that which we know"

PULMONARY ATRESIA

FEATURES

1 All blood flow to lungs is via the collaterals and bronchiolar arteries
2 Individual pulmonary shadows are small
3 "In a cyanotic like Fallot plus a continuous murmur (like P D A) most likely atresia of the pulmonary artery"
4 A loud single second sound (aortic) is present to the left of the pulmonary area
5 Unusual concavity of P A area
6 Small isolated shadows in the upper right mediastinal area and about hila of lungs
7 May have T of F with unilateral pulmonary atresia

PULMONARY STENOSIS WITH TRICUSPID ATRESIA AND UNDERDEVELOPED RIGHT VENTRICLE

FEATURES

1 Increasing cyanosis from birth
2 A systolic murmur is present over the pulmonic area
3 A normal sized heart
4 Relative left axis deviation
5 Left ventricular enlargement
6 Right ventricle is small

EBSTEIN'S ANOMALY OF TRICUSPID VALVE

1 210 000 of live births

FEATURES

A great deformity of the valves (except anterior) is present and these are placed downward into the ventricle

1 The atrium is greatly dilated causing an enormous cham ber
2 No signs of regurgitation
3 The patient is distinctly disabled A poor cardiac out put is noted
4 He is flushed or red faced with cyanosis of the lips
5 Dyspnoea or fatigue is noted
6 Cyanosis upon exertion is present
7 Syncope is noted in a few Headaches are present in a number A few may have seizures of petit mal type
8 Paroxysms of tachycardia are present
9 There is a systolic murmur and thrill (diffuse) Diastolic murmur is present in a few cases If tricuspid insufficiency is present a scratchy sound in early diastole is frequent (70%) A large quiet heart on palpation is characteristic of T I
10 Cardiomegaly is characteristic with Ebstein s anomaly
11 A split first sound is noted
12 Some have clubbing

(Refer to Kjellberg s book)

TYPES OF PULMONARY VENOUS CONNECTION

I PARTIAL

A Anomalous connection only to the left atrium
  (a) With normal lobular venous pattern
  (b) With abnormal lobular venous pattern

B Connection both of the right atrium or tributary veins and to the left atrium
  (a) With stenotic left atrial connection
  (b) With normal left atrial connection
  (c) With normal left atrial connection, but with mitral or aortic atresia

C Connections only to the right atrium or tributary veins
  (a) Without major intracardiac defects
  (b) With major intracardiac defects

D Connection between a pulmonary artery and the left atrium

E To S V C or I V C

F There is a very close association of A S D secundum with pulmonary venous anomalies

II TOTAL

Note

(1) Figure 8 shape in x rays
(2) Engorged pulmonary vessels
(3) In the lateral view the anterior mediastinum is remarkably clear
(4) Marked activity of the heart and the pulmonary vessels in fluoroscopy There may be central cyanosis
(5) Angiograms show filling defect in S V C

MAY HAVE
(1) Persisting left S V C to the coronary sinus right atrium right S V C or to the portal vein
(2) The pulmonary veins may pass into the right atrium or the coronary sinus
(3) Total right P V drainage into I V C
(4) Many combinations have been found and should be looked for

CLINICAL FEATURES OF ANOMALOUS PULMONARY VEINS
Malformation may cause early death
Venous and arterial blood are mixed in the heart
If no communication between the atria death occurs early
(1) A precordial bulge is present
(2) A parasternal "lift" on palpation of the area is readily noted
(3) A systolic murmur is present over the pulmonic area
(4) A wide mediastinum
(5) The right ventricle is hypertrophied
(6) Incomplete B B block is present
(7) A left to right shunt
(8) Marked lung vascularity

"Adventure on for from the littlest clue
Have come whatever man ever knew"

THE EISENMENGER COMPLEX
(E C) (8%)

(Victor Eisenmenger 1864 1932)

(Kindly refer to Dr Paul Wood's paper on the Complex and the Syndrome Brit M J May 1958)

CONSISTS OF TEN DIFFERENT ANOMALIES

FEATURES

1 Haemoptosis is prone to occur
2 The heart may be enlarged
3 *A systolic murmur and thrill (harsh) are present at the* base and are transmitted through the arterial tree
4 Aortic insufficiency may be a common finding The pul monary artery flow may be diminished
5 Arrythemias +
6 The second pulmonary sound is accentuated
7 The pulmonary blood flow is not diminshed The pul monary artery pressure is raised
8 Right to left shunt
9 Cyanosis later than T of F May be absent in infant, later persistent
10 A normal pulmonary valve The P A is dilated and a palpable impulse is present
11 There is slight or no clubbing *There is no squatting as* in T of F
12 Hoarseness and brassy cough +
13 *Sabot shaped heart when the P A is large* We must D D from Lutembacher The pulmonary arc is convex

and prominent in Eisenmenger s complex Hilar pulsations are to be noted

14 E C G or + Right axis deviation and a large biphasic Q R S complex is noted
15 The arterial blood O is important Using 70% diodrast shows dye in the aorta as soon as it enters the right ventricle (because of the overriding)
16 The aorta is dextroposed
17 A high ventricular defect
18 Abnormal aortic valve (frequent)
19 A thick right ventricular wall
20 Coronary insufficiency or +

Differential Diagnosis From Eisenmenger s (in absence of cyanosis) The following lesions should be considered

1 Malade de Roger and high V S D (The murmur is not transmitted)
2 Aortic and subaortic stenosis (normal pulse pressure)
3 Lutembacher (Get rheumatic fever history—mitral)
4 R H diseases (rheumatic heart)
5 T of F
6 Ayerza s disease and pulmonary hypertension
7 Pulmonary alveolar fibrosis (Rich syndrome)

We May See Evidence Of The Eisenmenger s Syndrome In

1 P D A
2 A P window
3 I V S D
4 I A S D and total anomalous pul V drainage
5 Single ventricle
6 Ostium primum
7 Transposition and I V S D
8 Single atrium
9 A S D

The cause of the main pulmonary dilatation is more likely to be due to the high pressure rather than to the flow between the two circulations

One gets endarteritis fibrosa in the lungs and thrombo obstructive lesions—right to left shunt and cyanosis later

---

"I hold that man is in the right who is most closely in league with the future"

TRANSPOSITION OF THE GREAT VESSELS (T G V)

(Karl Rokitansky 1804 1878)

FEATURES

1 Two separate circulations (like foetal life)
2 The Aorta is from the right ventricle The pulmonary artery is from the left ventricle
3 Severe cyanosis Only 10% of the pulmonary flow may pass into systemic circuit
Therefore Circulation 10 times greater than normal
Therefore Large heart progressively
4 Variable systolic murmurs
5 Right ventricular hypertrophy
6 The second sound over the pulmonary area is greatly ac centuated
7 The aorta is anterior to the pulmonary trunk
8 I V S D and P D A are present in a number of these

"Death! The spear that strikes but once"

INTERAURICULAR SEPTAL DEFECT (I A S D)

17% 1 13 500

TYPES

I UNCOMPLICATED

1 Patent foramen ovale No shunt
2 Complete absence of septum
3 A persistent ostium primum Left to right
4 *A persistent ostium secundum Left to right*
5 Localized single or multiple defects Left to right

II COMPLICATED

1 Atrioventricular communis Left to right
2 P F O with
 (a) A pulmonary stenosis with right to left shunt
 (b) Mitral stenosis with left to right shunt
 (c) Ebstein s tricuspid defect with a right to left shunt
 (d) Total anomalous pulmonary veins with right to left shunt
 (e) The 2nd component in the second heart sound may be widely separated during inspiration Note the large S V C and anomalous P V with this finding
3 I A S D (I-C D or E above) with
 (a) *A pulmonary stenosis with a right to left shunt*
 (b) A mitral stenosis with a left to right shunt
 (c) Ebstein s malformation and a right to left shunt
 (d) Tricuspid atresia with a right to left shunt
 (e) Transposition of great vessels with a right to left shunt

DIFFERENTIAL DIAGNOSES The following should be considered

1 Rheumatic fever
2 Left to right shunts (P D A I V S D with anom veins)
3 Pulmonary hypertension from other causes
4 I A S D with mitral stenosis indicates Lutembacher
5 I V S D
6 Aortic pulmonary "window
7 In ostium primum one gets early symptoms and pulmonary hypertension The M V and the T V are involved Pan systolic murmur is noted There is a left axis deviation

FEATURES

1 There is a predominent female sex incidence
2 There *may not be any symptoms* until late
3 A feeding problem is usually encountered in infancy
4 The child s body is of the slender type and is often under developed Rheumatic fever affects them more frequently than normal children
5 The skin is frequently translucent
6 Marfan s syndrome may be associated with an I A S D in 15% of cases
7 Respirogenic changes occur in youth as dyspnoea on ex ertion sighing fatigue and rapid respirations
8 Chest pain may be due to coronary insufficiency vascular dilatation or bronchial causes
9 A tapping apex beat or a tumultuous one due to over filling of the right ventricle is present There is a pre cordial bulge of the costal cartilages
10 There is a palpable pulsation in the out flow track
11 Cardiomegaly due to right auricle enlargement
12 Cervical venous pulsations are present when the normal child is recumbent but a wave is noted more distinctly
13 An hepatomegaly may be noted in failure of the heart It is insidious so that the hepatomegaly may lead to the diagnosis of longstanding heart failure

14 Cyanosis occurs in complicated cases only Here there may be added congenital lesions
15 A pulmonary systolic murmur with or without a thrill
16 A wide split second heart sound A thrill in 50% of cases
17 A diastolic murmur at the apex in $\frac{1}{3}$ cases A systolic murmur over the pulmonary artery area
18 A right bundle branch block is present in 95% of cases
19 In the E C G there is right axis deviation A large peaked P_2 in the E C G indicates an enlarged right auricle
20 Hilar pulsations are present
21 The aorta is small
22 A gross dilatation of the pulmonary artery
23 Pulmonary engorgement with repeated pneumonias" These are not inflammatory but due to alveolar congestion by blood
24 Pulmonary pressure may rise while the systemic blood pressure is normal
25 Globular shaped heart as seen in x ray
26 An enlarged right auricle and right ventricle
27 Pulmonary plethora Look for mitral stenosis in all cases
28 A left to right shunt Cyanosis is present when right to left shunt
29 Congestive failure may be early or late
30 Unexpected sudden death may occur at any age in infants with I A S D

INTERVENTRICULAR SEPTAL DEFECT (I V S D)

(12%) 1 4 000

A LEFT TO RIGHT SHUNT

COMMON

FEATURES

1 Symptoms may be absent until late Acute bacterial endocarditis is common
2 Growth may be restricted if a large I V S D is present Polydactylism and syndactylism are commonly noted (15%)
3 Cyanosis is absent Clubbing may occur if pulmonary hypertension is present
4 Dyspnoea is noted if a large I V S D varies by degree of defect
5 A loud rasping systolic murmur and thrill over the 3rd and 4th I C S and middle sternum "It spreads like a ripple on the water after a stone is thrown"
6 A normal sized heart if the defect is not large
7 The pulmonary artery is enlarged and it pulsates
8 A normal blood pressure in the absence of hypertensive lesions as C of A
9 A bulge may be present over the precordial region
10 A thrusting apex beat is noted in I V S D
11 The second heart sound at the pulmonary area is split A pulmonary diastolic murmur is present when the pulmon ary pressure is high in a small proportion of cases

12 A functional mitral diastolic murmur is present in 50% of cases
13 A pulmonary plethora is noted in the x ray film The chambers are often normal in size
14 The E C G shows a left and right ventricular hyper trophy but this may occur if pulmonary hypertension is present
15 A high oxyhaemoglobin level is found in the right ventricle in the presence of this left to right shunt This is noted by catheterization
16 The defect is high and between the aortic and tricuspid valves (90%)
17 Tricuspid incompetence may be present
18 A right interventricular systolic pressure equal to the pressure in the systemic arteries is presumptive evidence of a large I V S D
19 Aortic regurgitation may be associated with I V S D
20 Catheterization shows the location of defect Pressure and blood gas changes are noted Oxygenated blood is found in the right ventricle and indicates left to right shunt
21 Cardioangiograms may localize the defect Here one may see a second filling of the R V from the L V

DIFFERENTIAL DIAGNOSIS OF I V S D

1 Mild P V stenosis with a normal aortic root (higher site of defect)
2 Mild infundibular stenosis at aortic root (delayed P valve closure is noted)
3 Acyanotic examples of T of F
4 Mild aortic or subaortic stenosis (short systolic murmur)
5 Mild I A S D but heart contour is different
6 P D A A continuous murmur and is not systolic and alone as in I V S D
7 Haemic murmur

8 A high ventricular septal defect is not a true mal de Roger s

9 Pectus excavatum may cause a mechanical systolic mur mur over the same area

MITRAL ATRESIA (M A)

(Jean Baptiste De Senac 1693 1770)

1 Cyanosis from birth (decreased pulmonary flow)
Note (a) Ductus is closed
(b) E C G There is right axis deviation
2 Severe disability Early death unless attended to surgically
3 Precordial systolic murmur
4 The left ventricle is small The right ventricle shows hypertrophy
5 The great vessels may be transposed
6 A S D left to right shunt
7 Decreased pulmonary flow Due to high pressure in L A
8 Patent foramen ovale
9 There is a cor triloculare biatriatum with A V mixing of blood in the heart
10 *Less common than tricuspid atresia*
11 The aorta arises anteriorly and is narrower than the P A
12 Aortic valve atresia may be present

"A number of lesions in the adult heart are con genital in origin but missed or not looked for at a younger age"

MITRAL STENOSIS (Congenital)

1 Like acquired in symptoms but many variations
2 It is associated with I A S D in a number of cases Always search for it

For a full study of the subject kindly refer to the following detailed articles

1 *Congenital Mitral Stenosis*
By Charlotte Trenencz M D
Circulation Vol IX No 2 February 1954
2 *Congenital Anomalies of the Mitral Valve*
by John T Prior M D
American Heart Journal Vol 46 No 5 November 1953
3 *Congenital Mitral Stenosis*
By Henry Swan M D
American Heart Journal Vol 38 No 6 December 1949
4 *Congenital Mitral Stenosis*
By Robert W Cooley M D
Circulation Vol 76 No 4 October 1956
5 Refer also to more recent literature on this lesion

"Knowledge knows no frontiers"

LUTEMBACHER SYNDROME (L.S)

(Not common)

FEATURES

1 I A S D with mitral stenosis
2 Signs are those of these lesions separately
3 A large pulmonary artery is present
4 Jugular pressure is raised
5 Pressure gradient across the valve in I A S D generally means the presence of mitral stenosis
6 Incidence of stenosis is 5% with I A S D
7 In treatment one should open the mitral valve before closing the I A S D

COR TRILOCULARE BIATRIATUM

(Three chambered heart) (2 A plus 1 Vent)

(Thomas B Peacock 1812 1882)

1 Cyanosis is early mild and transient
2 A systolic murmur is heard over the entire precordium
3 X ray shows an unusual globular heart
4 Transposition of the aorta and the pulmonary artery is rarely seen
5 The O_2 saturation of the arterial blood is subnormal
6 The E C G shows high voltage A diphasic Q R S complex is present
7 The O saturation of the haemoglobin in the ventricle exceeds the haemoglobin of mixed venous blood (catheter)

"The reward of success will attend our efforts in so far as the heart is treated as one of the ordinary tissues of the body and the method of operation conforms to the fundamental principles of surgery"

SIR RUSSELL C BROCK
The 1958 Bradshaw Lecture

1883 Billroth s unwise remark

"The surgeon who should attempt the suture of the heart would lose the respect of his colleagues"

"Even though the heart is but 1 cm from the surface of the body it has taken us 2000 years to reach it"

TAUSSIG-BING COMPLEX (T B C) (Rarest) (INCOMPLETE TRANSPOSITION WITH BIVENTRICULAR ORIGIN OF PULMONARY ARTERY)

1. Cyanosis
2. A transposed aorta
3. A large pulmonary artery from the right ventricle which overrides the ventricular septum
4. A high V S D

DEXTROCARDIA

1 The primitive cardiac loop swings in the reverse direction to the normal
2 The course of the circulation is the mirror image of the normal Situs inversus of viscera is always present
3 Asymptomatic unless other lesions are associated with it
4 The E C G is pathognomonic
5 May have a left aortic arch with cyanosis
6 Bronchiectasis is frequently associated with dextrocardia due to pressure of the abnormal position of the heart on the bronchi
7 Auscultation and x rays pick it up

Differential Diagnosis

1 A displaced heart from atelectasis pneumothorax or congenital absence of the right lung
2 Incomplete rotation of the heart
3 Right ventricular hernia or a pericardial cyst

TRICUSPID ATRESIA (T A)

(William Harvey 1578 1659)

FEATURES

1 Cyanosis from birth
2 The I V C and S V C syndromes are present The venous pressures are elevated
3 Severe spontaneous dyspnoeic attacks may occur
4 A basal systolic murmur is present
5 If a picture of T of F is present but hypertrophy of the left ventricle and E C G changes it is not likely a T A
6 Eighty per cent have an associated pulmonary stenosis
7 There is no communication between the right auricle and the right ventricle
8 The P A is smaller than the aorta
9 There is a configuration of a Dutch shoe in the contour of the heart
10 The E C G shows left axis deviation
11 A patent F ovale is present and the ductus arteriosus is not patent
12 The pulmonary trunk may arise from a small R V and can be reached only via the I V S D
13 Cardioangiograms are useful here

PERSISTENT COMMON ATRIOVENTRICULAR CANAL

(Franklin Paine Mall 1862 1917)

FEATURES

1 A V canal is undivided and guided by a single valve of a large posterior cusp and small lateral cusp
2 An I A S D is present
3 Variable cyanosis—mistaken for I A S D (primary)
4 A systolic murmur is heard over the mid precordium and apex
5 A left and right ventricular hypertrophy is present
6 The E C G shows high voltage Biphasic Q R S complexes are noted

BLAND-WHITE-GARLAND SYNDROME

FEATURES

1 Attacks of dyspnoea, cyanosis and pallor
2 Coronary ischaemic pain drawing up of the legs and extension of the neck during and after feeding May have a soft blowing murmur
3 The left auricle is greatly enlarged Feeding problems occur after two months
4 There is evidence of antero-lateral infarction in E C G in the infant
5 The left coronary artery arises from the pulmonary artery
6 Sudden death may occur at 2 13 months
7 Should differentiate from subendocardial fibroelastosis congenital aortic stenosis and Fiedler s myocarditis

PERSISTENT TRUNCUS ARTERIOSUS

(Sir Arthur Keith 1866-   )

FEATURES

1 A single arterial vessel (aorta and pulmonary artery are one)
2 Arises from both ventricles
3 A short septum in the truncus divides it into a short aorta and pulmonary artery
4 Cyanosis moderately severe
5 X ray—right and left ventricle enlarged
6 Hilar shadows increased when cyanosed
7 E C G shows right axis deviation
8 O_2 saturation is below normal
9 No P D A

BLAND-WHITE-GARLAND SYNDROME

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CONGENITAL PULMONARY ARTERIOVENOUS FISTULAE

A manifestation of hereditary haemorrhagic telangiectasis (redundant Osler Weber disease) in percentage of cases

THE FOLLOWING FEATURES MAY BE NOTED

1 Cyanosis and clubbing of fingers and toes There is dyspnoea on exertion
2 Haemoptosis and epistaxis
3 Chest pains thoracoepigastric pain in adults due to pulmonary hypertension
4 A continuous hum or systolic murmur with extraventricular systoles is frequently detected
5 No cardiac enlargement.
6 Forty percent have telangiectasis elsewhere in the body E g Maffucci s syndrome
7 The red blood counts are increased (polycythaemia)
8 The blood viscosity and blood volume are increased
9 The haematocrit is greatly increased
10 The O_2 saturation is decreased to 70%
11 X ray findings of opacities (discs) or grape like shadows in the lung field
12 The incidence of S A B E increases
13 If cerebral thrombosis occurs due to increased anoxia and viscosity then headaches vertigo convulsions and paresis may occur
14 Variation in size of the lung shadow with Valsava s experiment. Must differentiate from Arrilaga Ayerza s dis

COR BILOCULARE
(TWO CHAMBER HEART)

(William Hunter 1718-1783)

Features

1. Infant death is characteristic of this lesion
2. No murmurs
3. Early and progressive cyanosis
4. A single atrium
5. A single ventricle
6. The P A lies behind the aorta
7. The flow to the lungs is via the P D A
8. *Most primitive* heart
9. Dextrocardia *often is associated*
10. Globular enlargement of the heart
11. The atrium receives blood from the lungs via the pulmonary veins
12. Peripheral blood via the I V C and a right and a left S V C are present.

CONGENITAL PULMONARY ARTERIOVENOUS FISTULAE

A manifestation of hereditary haemorrhagic telangiectasis (redundant Osler Weber disease) in percentage of cases

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1 Cyanosis and clubbing of fingers and toes There is dy spnoea on exertion
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13 If cerebral thrombosis occurs due to increased anoxia and viscosity then headaches vertigo convulsions and paresis may occur
14 Variation in size of the lung shadow with Valsava s ex periment Must differentiate from Arrilaga Ayerza s dis

ease, Tetralogy of Fallot and Eisenmenger malformation
15 May associate Maffucci's syndrome and haemangiomata with A V fistula and hemihypertrophy of the limbs (See Wm Beans work of Iowa City)

Let us not mistake extracardiac lesions as coelomic cysts pulmonary A V fistulae or pericardial effusions for primary cardiac lesions"

The Principle Clinical Findings In Cyanotic Heart Lesions

J W Brown

Signs and Symptoms	*Tet al gy of Fall t*	*Eisenmenger*	*Pul nonary St no si F O Ial nt*	*Tr cuspid Atre a*	*Tran posit on of Great V sels*	*Persistent Trun cus A teri sus*	*S ngle Ventricle*
CYANOSIS	Early and marked	Late and slight	Early Prog es s v ly mark d	Marked	Marked	Marked	Marked
CLUBBING	Ma ke l	Slight	Sl ght to ma ked	Marked	Marked	Marked	Marked
POLYCYTHEMIA	Severe	Later stages	Severe	Severe	Severe	Severe	Severe
SYMPTOMS	Severe	Ch ldhood	Present early	Present early	Present early	Present and severe	Severe and early
PHYSICAL CAPACITY	Grossly impaired	Little	Se ere	Grossly	Grossly	Grossly	Gro sly
DYSPNOEA	Se ere Squats	Some	Severe	Severe Squats	Severe	Severe	Severe
SYSTOLIC MURMUR	May be absent In pul area	Pulmonary area	Pulmonary area	Absent	Often absent with I V S D	Present or absent	Present or absent
DIASTOLIC MURMUR	Rarely present	Frequ nt from aortic or pul incompetence	Occas onally	Absent	Absent	Occasionally	Absent
PULMONARY SECOND SOUND	*Unaltered often*	*Accentuated*	*Dimini hed*	*Loud and not reduplicated*	*Unaltered*	*Loud and pure*	*Unaltered*
X RAY ENLARGEMENT	Normal	Slight to moderate	Sl ght enlargement	Often present	Enlargement	Gross	Moderate
PULMONARY ARC	Concave	Convex and up	Convex and up	Concave	Concave	Concave in late childhood	Early slight convexity
HILAR SHADOWS	Small	Normal	Normal	Small	Normal	Small	Small
HILAR PULSATION	Nil	Present	Present	Absent	Present	Absent	Absent
PULMONARY CONGESTION	Nil	May be present	None	None	Present	None	None
E C G R AXIS	Marked	Marked	Marked	Left axis	Rt axis Oc casional normal	Rt Axis	Rt. Lt. Axis
P WAVES	Tall & peaked	Enlarged	Enlarged	Enlarged	Enlarged	Enlarged	Enlarged
CIRCUL TIME	*Shortened*	*Shortened*	*Unaltered*	*Unaltered*	*Unaltered*	— — — —	— — —

WHEN SHOULD AN OPERATION TAKE PLACE (LAM)

Diagnosis	*Type of Operation*	*May be Done*	*Should be Done*	*Youngest Successful*
AORTIC RING	Division	Soon as diagnosed	Soon as diagnosed	3 weeks
PATENT DUCTUS ARTERIOSUS	Ligation or division	Soon as diagnosed	Soon as diagnosed	6 weeks
COARCTATION	Resection	As early as possible	2 years plus or earlier if necessary	16 months
VALVULAR PULMONARY STENOSIS	Valulotomy	As early as in trouble	As early as in trouble	7 months
TETRALOGY OF FALLOT	Potts or Blalocks	As early as in trouble	2 years plus or earlier if necessary	9 months
TRICUSPID ATRESIA	Blalocks or Potts	As early as in trouble	After 15 18 *months* if possible	5 *months*
I A.S.D	Closure	With symptoms and pulmonary hypertension	Enlarging auricle	6 months
I V S.D	Closure	After S A B E	Symptoms and Lt Rt. shunt	6 months

CARDIAC ARREST
—THE DIAGNOSIS AND TREATMENT—

1 The heart is the most efficient pump in the world If it stops we must restore the beat "within three minutes" Each year it beats 37 879 700 times!!

2 Signs of cardiac arrest
After a period of slowing and a lowering blood pressure there are the following
(a) No pulse
(b) No blood pressure
(c) Dilated pupils
When did the heart stop beating? We can only know the answer from the anaesthetist in charge It is he who is the best "monitor" An alert anaesthetist is the best monitor"

3 Cardiac arrest is like turning off the ignition in a good car In C A something has been turned off

4 *The heart never ceases to beat except for an adequate* reason Its cause can generally be found on review

5 It will be malpractice by 2000 A D not to attempt restoration of the heart beat when cardiac arrest occurs in an otherwise healthy body A number have been restored following coronary infarction and should be attempted in such cases in the absence of severe arteriosclerosis or old age

6 Cardiac arrest may occur anywhere at anytime during examination or performing even a minor operation upon a patient

7 All who practice medicine should be able to massage an arrested heart within three minutes of its cessation

One should never be criticized for trying to save a life

8 Every room in the Emergency Ward and the O R should have a small tin case hanging on the wall containing the following
 (a) Intratracheal tube (adult and child)
 (b) A scalpel
 (c) A 20 c c syringe with a long fine needle
 (d) Calcium chloride—5% ampule
 (e) Procaine—1% ampule
 (f) Adrenaline—1 10 000 ampule
 Oxygen should always be available

9 Every O R should have a ready defibrillator

10 The brain MUST have blood loaded with O_2 to survive beyond three minutes
 Therefore remember

 PUMP + OXYGEN + BRAIN + HEART—
 before three minutes

11 Aetiology of Cardiac Arrest
 (a) Poor preparation of the patient—physically and mentally
 (b) O_2 lack in the brain and heart
 (c) CO_2 increase in lungs and circulation
 (d) Electrolytic imbalance and excess of drugs
 (e) Reflex stimulation of vagus nerve
 (f) Lowered blood pressure with anoxia
 (g) Lack of careful observation or "watchful expectancy" on the part of the attendant

12 Rarely do signs come on suddenly" They appear "sudden" because the pulse and the B P have not been watched constantly

13 Potassium citrate and acetylcholine cause cardiac arrest

14 Calcium chloride controls the rhythm of the heart beat

PREVENTION OF CARDIAC ARREST

1 Make the patient safe for surgery
2 Think of its possibility during any examination
   E g Carotid body Before and during any operation no matter how minor
3 Watch for it. The best "monitor is an alert anaesthetist
4 Use of an amplifying stethoscope E C G
   Electric monitor or a stethoscope strapped over the heart where possible during the operation
5 The use of digitalis pre operatively in bad risk cases
6 Adequate O_2 and CO_2 control
7 Prevent hypotension
   "Treat the tissues as though they were cobwebs" thus prevent shock
8 Increase pulmonary reserve by respiratory exercises
9 Keep bronchi and bronchioles clear of secretions
   Allow O_2 to reach the alveolar sacs
10 Select the safest" anaesthetic
11 Adequate level of sedation
12 Careful consideration of added drugs during prolonged operations An accumulative effect may occur
13 Use of atropine (prevents bradycardia)
14 Keep the fingers on the pulse instead of "crossing the fingers! Avoid constricting arm bands or twisted neck
15 Avoid compressing a sensitive carotid body
16 Careful and gradual induction of anesthesia
17 Adequate airway (size and depth)
18 Continuous observation of pulse and B P A heart always slows before stopping this is the time to awaken to the danger of arrest Stay by the patient
   When did the heart stop beating? The answer gives the prognosis
19 Anaesthetize the trachea before intubation

20 Care with hyperventilation with O_2 at end of operation Too sudden release of built up CO_2 by O_2 is dangerous
21 Care with positioning and posturing the patient If in slight Trendelengerg on the operating table then the stretcher and bed must be ready in this position before moving the patient

IF THE HEART HAS STOPPED BEATING

"It is TIME teamwork massage oxygen and tena city all together"

1 Do not question it or stand around
2 Do not listen for a "faint beat, that faint" beat has al ready passed!! It's later than you think!
3 Do not wait for an E C G or B P apparatus
4 Do not ask for gloves or gown but ONLY for a scalpel knife or blade

"You can clean up infection but you cannot clean up death"

5 Do not use adrenalin into an unseen heart
6 Do not use mechanical respiration
It is a pump you need the pump is your hand Use it immediately
7 Do not force O_2 into the trachea, the blood must be moving before it can reach the brain--give O_2 and massage to gether
8 Do not dilate the anal sphincter
9 Do not transfuse or increase I V until massage has started and O_2 is reaching the lungs and brain If C A is from haemorrhage, then force *blood quickly*
10 Do not go through or massage diaphragm
11 Do NOT WAIT A SECOND or waste time "hoping" it is not C A

BUT AT ONCE--take courage and get to the heart IMMEDIATELY while the following are being done (at the same time)

1 Note time by clock (Have a time-caller)

2 Stop anaesthetic at once
3 Tip table down 15° Give O_2 via the tube or mask.
4 Open 5th left interspace wide (in 6 seconds)
5 Spread ribs with hands later with spreaders
6 Open pericardium Note heart.
7 If not fibrillating (only 10% do) massage with one or both hands Compress and relax 40 - 50 times per minute Steady and evenly B P will now appear
8 If fibrillating, apply defibrillator If not one about use an electric current to heart or rapid needle pricks into heart muscle might benefit. But, MASSAGE always keep the brain oxygenated until fibrillation is controlled and the heart beat restored.
9 The heart will always respond to massage but the brain cannot go without O_2 longer than three minutes
10 If the heart is flabby and irregular inject one of the following and massage
   (a) 10 cc of 5% calcium chloride into left ventricle
   (b) 3 cc of 1 10 000 Adrenalin
   (c) 9.5 cc 1% procaine hydrochloride and .5 c c adrenalin into L V
   These may be repeated with care
11 Massage and observe but massage
12 Massage until rigor mortis sets in if necessary This will not set in if we massage early and give O_2 before three minutes of cessation
13 Bleeding from the wound is a good sign that blood is reaching the brain also
14 When all is restored, then attend to closure and observe the brain and kidney post-operatively for any damage due to cardiac arrest May use hypothermia for several days

OMEGA

If this little book causes you to see what you look for or stimulates you to learn what you do not know in the diagnosis of congenital heart disease, if it causes you to see the "Great Path of Duty" in Medicine—correct diagnosing and to follow it more bravely, more faithfully and more successfully if it will stimulate the graduate medical student to enter the field of cardiac surgery or cardiology in which field so much more can be accomplished *it then will have* accomplished the chief object that prompted the author to write "Toward the *Diagnosis of* Congenital Heart *Disease*"